

**EBSTEIN'S ANOMALY WITH
WOLFF-PARKINSON-WHITE SYNDROME
AND FREQUENT REPETITIVE
TACHYARRHYTHMIC EPISODES
BASAL12-LEADS ECG:**

- 1. Pseudo Brugada type 1 ECG pattern***
- 2. Pseudo J-Osborn wave and***
- 3. Pseudo RBBB.***

**Case of Dr Raimundo Barbosa Barros Fortaleza – Ceará - Brazil
Final conclusions by Andrés Ricardo Pérez-Riera.**

CASE REPORT

These tracing belongs to a 46-year-old woman, carrier of Ebstein's anomaly, admitted in the emergency room with atypical chest discomfort and palpitations, without risk factors for coronary artery disease, normal biomarkers, and serial unaltered ECGs.

The first tracing is similar to the prior ones made over the last 7 years. This one was made immediately after cardioversion (there are no artifacts).

All the ECGs in her history show the same pattern.

She displays Wolff-Parkinson-White with frequent paroxysmal palpitations.

Five years ago ablation was proposed to her; however, the patient refused the procedure afraid of complications.

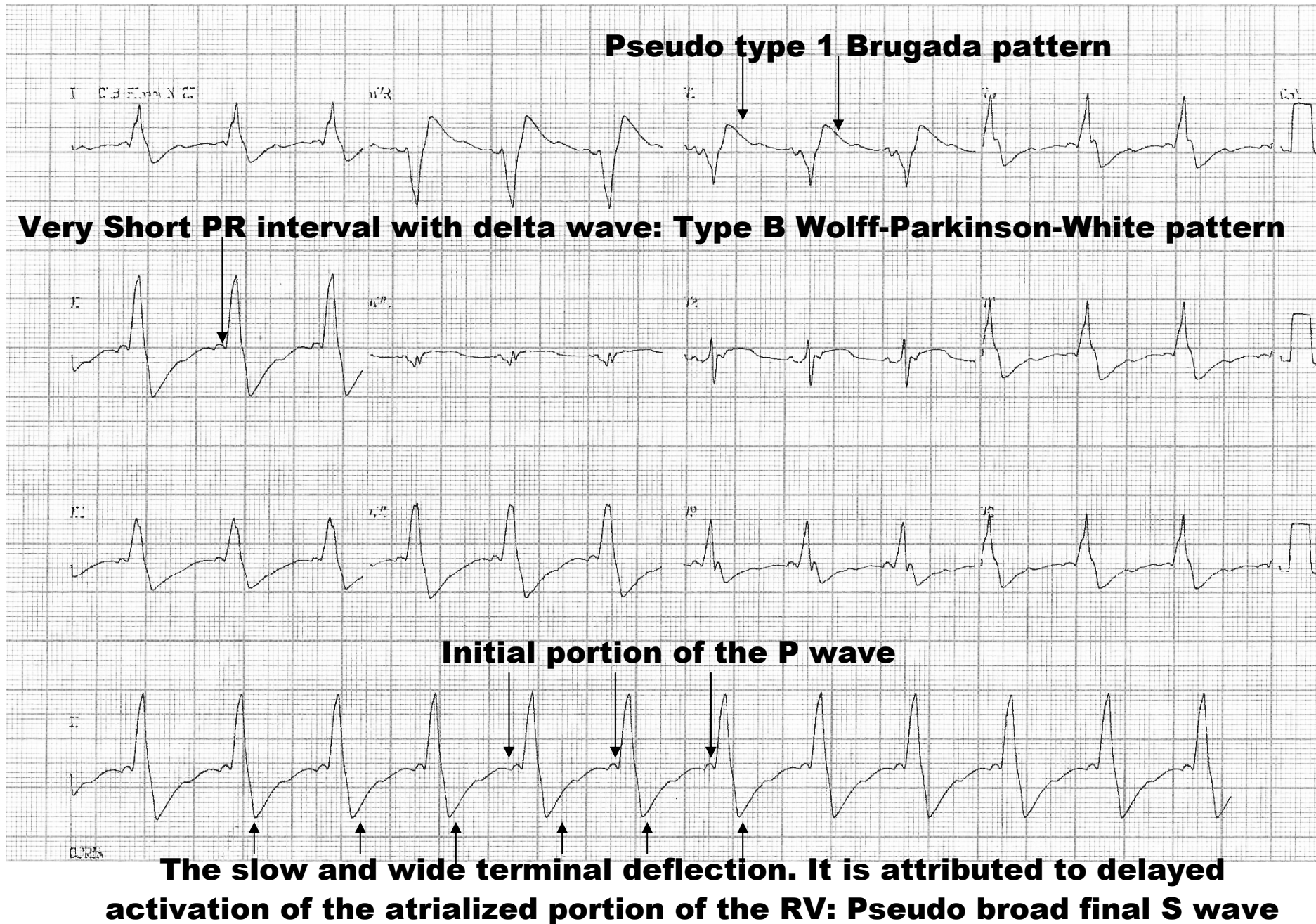
Question: Is this notch inside of QRS: is this a J wave? Is a conduction disorder? or is part of a giant wave P? What do the colleagues think? Already underwent one unsuccessful ablation

Estes traçados pertencem a uma mulher de 46 anos de idade, portadora de anomalia de Ebstein, admitida na sala de emergência com desconforto de torácico atípico e palpitações. Sem fatores de risco para doença arterial coronária, biomarcadores séricos normais, e ECGs seriados inalterados. O primeiro traçado é semelhante aos realizados periodicamente nos últimos 7 anos. Este foi realizado logo após cardioversão elétrica (não existe nenhum artefato). Todos os ECGs pesquisados na sua ficha da história clínica mostram-se com igual padrão. Portadora de síndrome de Wolff-Parkinson-White que lhe ocasiona palpitações paroxísticas frequentes.

Cinco anos atrás foi lhe proposto tratamento curativo dos eventos com aplicação de energia de radiofrequência que a paciente recusou com temor de complicações.

Pergunta: Este entalhe dentro do QRS é uma onda J? É um distúrbio dromótopo? É parte de uma onda P gigante? Que pensam os colegas. Já foi realizado aplicação de energia de radiofrequência sem sucesso.

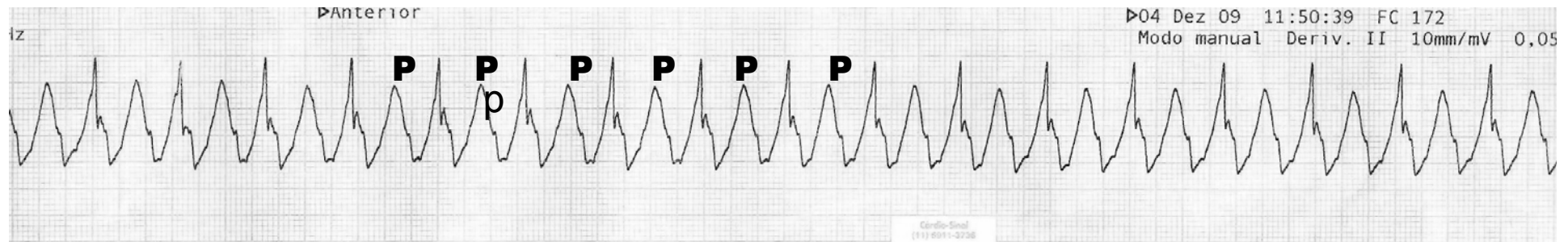
ECG PERFORMED IMMEDIATELY AFTER ELECTRICAL CARDIOVERSION



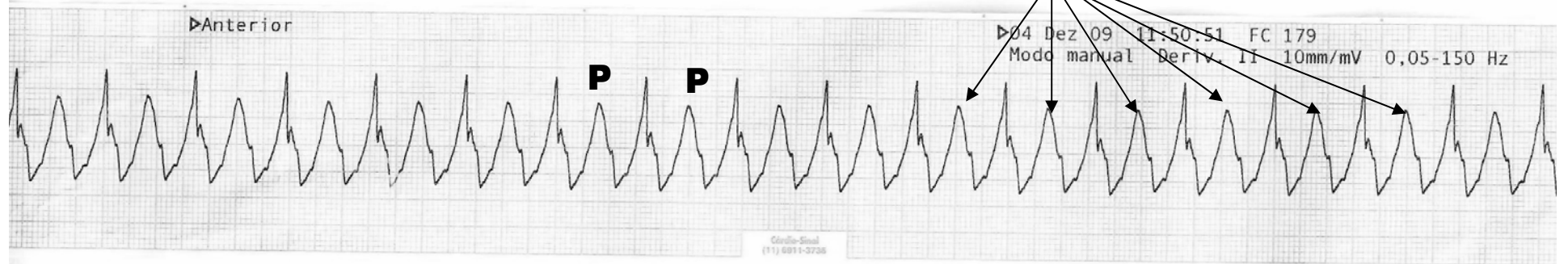
ECG DURING EVENT



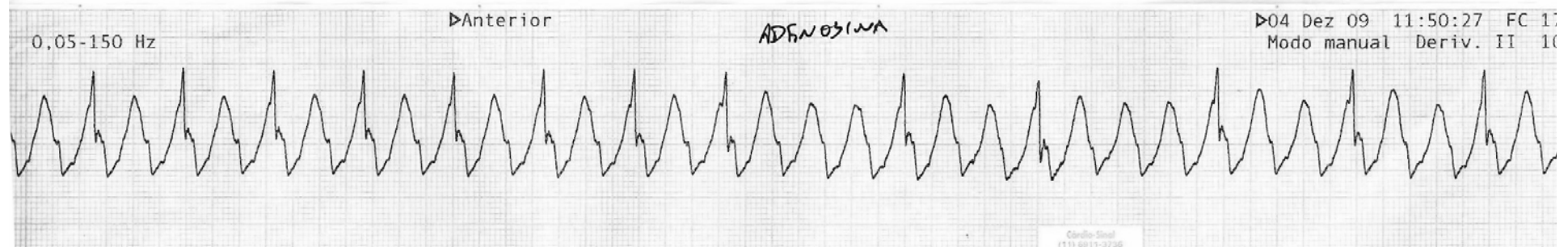
AFTER ADENOSINE (CONTINUOUS LEAD II)



Atrial Tachycardia with 2:1 AV block. Himalayan P waves.

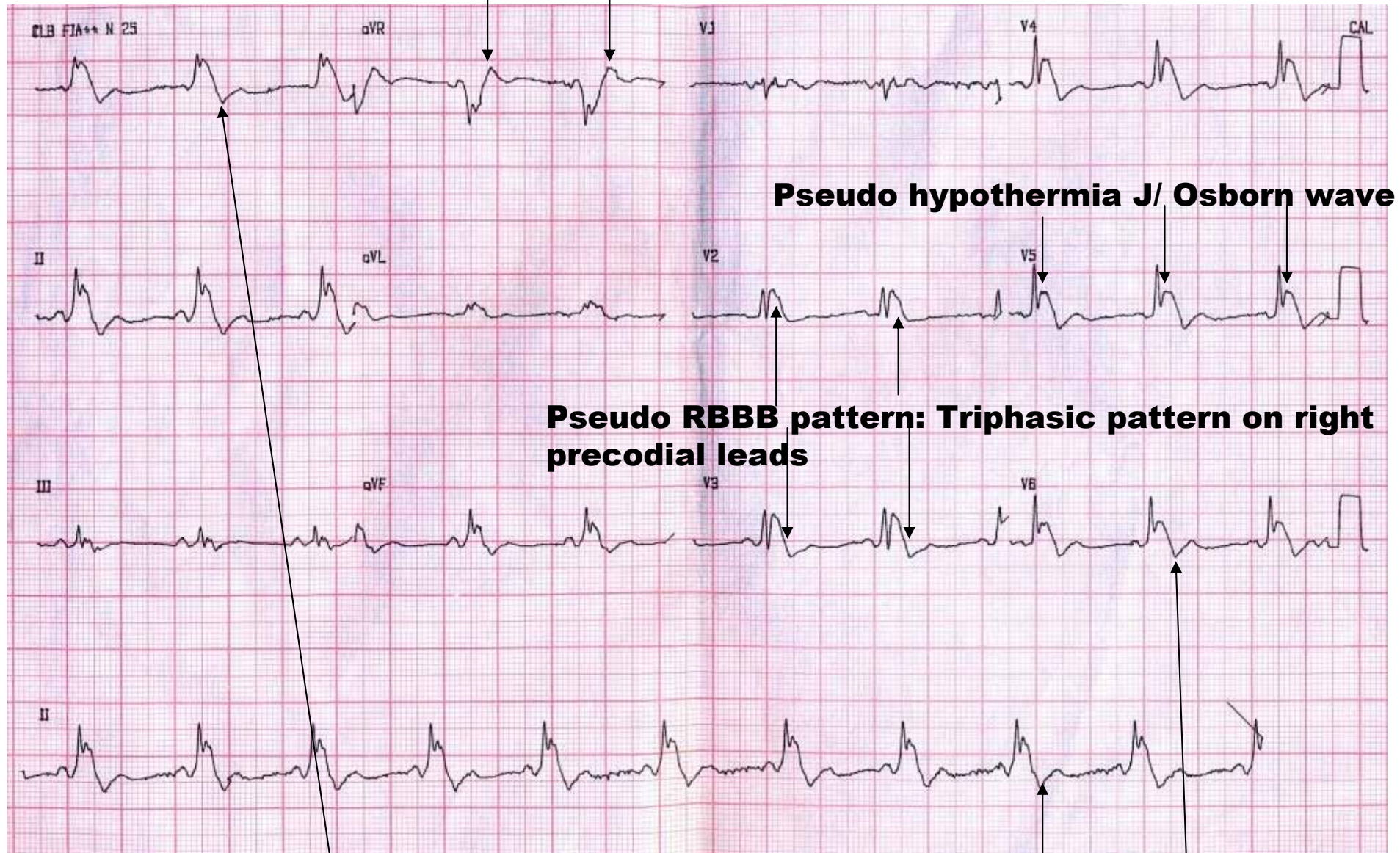


Taquicardia atrial com Bloqueio AV 2:1 com ondas P gigantes.



AFTER CARDIOVERSION

Pseudo broad final r' wave in aVR: Pseudo RBBB



Pseudo RBBB pattern: Wide pseudo final S wave on left leads I, V5-V6

Modo manual Deriv. II 10mm/mV 0,05-150 Hz



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Modo manual Deriv. II 10mm/mV 0,05-150 Hz ADENOSINE



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COLLEAGUES OPINIONS
OPINIÕES DOS COLEGAS

Para mi es una onda P en la derivación DII de todos los ECG de 12 derivaciones.
Tiene múltiples vías y por eso es difícil de ablacionar. Apesar de esto pero hay que insistir caso permanezca muy sintomática.

Pregunto: ¿porque emplearon adenosina?

Realmente me gustaria saber a que muesca usted se refiere?

For me it is a P wave in lead II of all 12-lead ECG.

She has multiple pathways and is therefore difficult to ablate. In spite of this, but if we must to insist if she remain highly symptomatic.

Wonder why adenosine was used?

I really want to know what you mean notch?

Para mim é uma onda P em DII de todos ECG de 12 derivações.

Tem múltiplas vias anômalas em paralelo e por isso é difícil de curar com a radiofrequência. Apesar disto, é necessário insistir se permanecer altamente sintomática.

Pergunto por que foi usado a adenosina?

Eu realmente gostaria saber a que entalhe você se refere?

Carlos Rodriguez MD crartuza@hotmail.com

This is a fairly common variant of Ebstein's anomaly. The slow terminal deflection is attributed to delayed activation of the atrialized portion of the RV which can be confirmed by intracardiac ECG. To my knowledge it has not been called J wave like in hypothermia.

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Esta é uma variante muito comum na anomalia de Ebstein. A deformação lenta terminal é atribuída a ativação retardada da porção atrializada do VD que pode ser confirmado pelo ECG intracavitário. Que seja de meu conhecimento não tem sido chamada de onda J-like como em hipotermia.

Esta es una variante bastante común de la anomalía de Ebstein. La deflexión terminal lenta se atribuye a la activación retardada de la parte atrializada del VD que puede ser confirmada por ECG intracardiaco. Que yo sepa no se ha llamado de onda J-like como en la hipotermia.

FINAL CONCLUSION

1. Typical ECG of Ebstein anomaly
2. Type B Wolff-Parkinson White Syndrome with APs located in the RV free wall (between the RA and the RV). The association with *WPW in ebstein's anomaly* occurs in 5 to 10% of the cases. There are authors that suggest a greater percentage of associated pre-excitation (25%). Finally, some authors think that type-B WPW associated to tachyarrhythmias is observed in more than 50% of cases¹. This is the congenital heart disease most associated to *WPW*.
3. Wide or broad QRS complex antidromic reciprocant macro-reentry tachycardia or Circus Movement Tachycardia with probably several right APs
4. Pseudo atypical Brugada ECG type 1 pattern during sinus rhythm
5. Pseudo Right Bundle Branch Block during sinus rhythm
6. Pseudo hypothermal J wave or Osborn wave during sinus rhythm
7. Atrial tachycardia with 2:1 AV block after adenosine
8. Himalayan P waves of Tausing. Ebstein's anomaly is a rare complex fascinating congenital anomaly with a broad pathologic-anatomical and clinical spectrum accounting for <1% of all congenital heart defects. Since its description in 1866, dramatic advances in diagnosis have been made. Very high P waves “like-Himalaya mountain”² are observed. (the Himalayan mountain system are the planet's highest peaks around the world's). The P wave is > 3mm (0.3 mV) in near 50% of cases³. Tall P waves (≥ 2.5 mm) are attributable to right atriomegaly.

1. Deal BJ, Keane JF, Gillette PC, Garson A Jr. Wolff-Parkinson-White syndrome and supraventricular tachycardia during infancy: management and follow-up. J Am Coll Cardiol. 1985; 5: 130-135.
2. Kaushik ML, Sharma M, Kashyap R. 'Himalayan' p wave. J Assoc Physicians India. 2007;55: 856.
3. Jaiyesimi F. Observations on the so-called non-specific electrocardiographic changes in endomyocardial fibrosis. East Afr Med J. 1982 Jan;59:56-69.

In Ebstein anomaly are frequently observed¹:

- 1. Prolonged PR interval (≥ 170 ms in childrens)**
- 2. Short PR interval if associated with WPW pattern($\approx 30\%$ of cases²)**
- 3. Bizarre low voltage right bundle branch block pattern with initial q wave in QRS complexes on right precordial leads V_1 and V_2**
- 4. Reduced amplitude of R-wave deflections in V_{3R} and V_1**
- 5. P-dextro-atriale and bizarre low RBBB without right ventricular overload almost certainly constitutes a pathognomonic finding in Ebstein's anomaly,**
- 6. Higher P waves and wider the QRS complexes are registered in more severe cases of Ebstein's anomaly of the tricuspid valve.**
- 7. High potential for developing arrhythmia, in the vast majority, of the tachycardia type:**
 - Atrial ectopic tachycardia,**
 - Atrial flutter**
 - Atrioventricular reentry tachycardia**
 - AV-nodal reentry tachycardia**
 - Atrial fibrillation and**
 - Ventricular tachyarrhythmias.**

Tendency towards multiple arrhythmogenic substrates in the single patient².

- 1. Blömer H. Electrocardiographic and phonocardiographic findings in Ebstein's anomaly with special regard to its severity. Med Klin. 1975; 70: 1175-1178.**
- 2. Hebe J. Ebstein's anomaly in adults. Arrhythmias: diagnosis and therapeutic approach. Thorac Cardiovasc Surg. 2000; 48: 214-219.**